Imaging features of small bowel and colorectal cancer in inflammatory bowel disease

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Learning Objectives

• To know the risk factors for developing small bowel cancer (SBC) and colorectal cancer (CRC) in patients with inflammatory bowel disease (IBD)

• To know the clinical and pathological features of CRC and SBC in patients with IBD

• To illustrate the imaging features of small bowel and colorectal malignancy in IBD

• To emphasize the difficulty in establishing a diagnosis
Background

Inflammatory Bowel Disease (IBD)
- Ulcerative Colitis (UC)
- Crohn’s Disease (CD)
- Indeterminate Colitis (IC)
Patients with IBD have **increased risk** for developing colorectal and small bowel cancers.

The **prevalence** of CRC in patients with UC is approximately **3.7%** overall and **5.4%** for those with pancolitis (1).

The prevalence of CRC in Crohn’s colitis is similar (2).

Patients with CD have **relative risk 28** for developing small-bowel cancer compared to the background population (3).

**No increased** risk was found in stomach and anal cancer in patients with IBD (4,5).
Risk factors for CRC in patients with IBD
Duration of Disease

The risk of CRC becomes greater than that of the general population after 8 to 10 years from the onset of disease (1).

The cumulative incidence of CRC is 2.5% after 20 years of IBD, 7.6% after 30 years and 10.8% after 40 years (7).
**UC:** the standardized incidence ratio (SIR) for CRC increase gradually from 1.7-fold in proctitis, 2.8-fold in left-sided colitis to **14.8-fold in pancolitis**, compared with age-matched population without UC (8).

**CD:** the risk of CRC is increased when the extent of the colic involvement is greater than one third(7).
Primary Sclerosing Cholangitis

The **concomitant** presence of **PSC** in IBD patients confers a high risk for developing colorectal cancer(9,10).

The cumulative incidence of CRC in UC patients was 33% at 20 years (10).

When **liver transplantation** is necessary, prophylactic colectomy should be considered(11).
• Young age at onset, when younger than 25, increases the risk of developing CRC (1).
• When onset is after 30 there is no increased risk of CRC.

• Family history of sporadic CRC increases twice the risk of CRC (13).

• The increased severity of inflammation correlates with increased frequency of dysplasia (7).
• Patients with longstanding quiescent colitis remain at risk for developing CRC.
Screening colonoscopy should begin in patients with IBD:

- **8-10 years** after the onset of IBD symptoms
- UC pancolitis or left-sided colitis
- Crohn’s colitis involving at least one third of the colon
- At onset of PSC if associated
Clinical and pathological features of colorectal cancer in IBD

Age at diagnosis
- 10 years earlier than in sporadic CRC
- UC: The mean age is 52 years (14)
- CD: The mean age is 54 years (15)

Anatomical Location
- Tumor occurs in area of macroscopic disease
- CD: Tumors occur in ileocaecal and rectosigmoid regions
- UC: From the rectum to the right-sided colon (15)

Histology
- Frequency of mucinous and signet ring cell tumor is higher than in the general population (15)
- Synchronous tumor locations
Risk factors for SBC in patients with Crohn’s Disease
The relative risk of small bowel carcinoma in CD seems to be 28.4 times higher compared to general population (3).
Essential factor

The mean duration of CD is 19 years (3).

Distal jejunal and ileal location (16).

When younger then 25 years at onset of CD.

Strictures
Chronic fistulous disease
Small bowel bypass loops.
Clinical and pathological features of SBC in CD

- **Age at diagnosis**: The median age of diagnosis is 48 years versus 65 in general population (16).

- **Anatomic location**: The highest incidence in the **distal jejunum and ileum**: area of macroscopic disease.

- **Histology**: Adenocarcinoma with signet ring cell is frequent: up to one third (17).
Patients and materials

The computerized medical record system Explore in the PACS at the Radiology department of the University Hospital of Nancy was used to identify patients with IBD and concomitant SBC or CRC.

The diagnosis of IBD, CRC and SBC were confirmed by clinical, imaging, endoscopic and histological criteria.

Only patients who had a scanner or magnetic resonance were accepted.

There were 15 patients with both, IBD and CRC-12 or SBC-3, between 2001 and 2009.
There were 12 patients with IBD and concomitant colorectal cancer. 

8 of them had Crohn’s disease and only 4 had Ulcerative Colitis.
75% of the patients with CRC had severe **pancolitis**.

40% of the CRC were located in the **left colon**.
42% of CRC were with Signet Ring Cell component. CRC with signet ring cell are only 1% of CRC in general population.

Histological aspect of colic adenocarcinoma with signet ring cell component. This is a signet ring cell pattern of adenocarcinoma in which the cells are filled with mucin vacuoles that push the nucleus to one side, as shown at the arrow.
Only 3 (25%) of CRC were discovered by screening colonoscopy. Five (42%) of CRC presented an occlusion. Three (25%) had worsening of the IBD and one had anemia.

In these 5 cases of CRC presenting as occlusion the pre operative diagnosis of neoplasia was not suspected. The imaging findings indicated an inflammatory benign stenosis.
39 year-old man with ileocecal CD from 26 years and PSC with liver transplantation 8 years ago. Refusal of screening colonoscopy. CT and MRI realized for worsening of the CD with inefficient medical treatment.

Asymmetric wall thickening of the right colon with a parietal mass. Hyperintense asymmetric mass in T2 weighted images related to hydric mucinous component. Poor and late enhancement after gadolinium IV administration relegated to the fibrous tumor. Pericoloic spiculations in the peritoneal fat and positive ganglia on histology.

Surgery- Mucinous adenocarinoma pT4N1M0
58 years old female with CD from 2 years and long history of digestive disorder. Pancolitis and difficult medical treatment, right-sided colon stenosis.

2 years later. Stenosis and right-sided colitis responsible for small bowel occlusion. The stenosis was present in 2007.

Ileocaecal surgical resection: adenocarcinoma with signet ring cell: T4N2M0.

Same patient 2 years later: 60 years old. Small bowel occlusion resistant to medical treatment since 2 months and lost of 2 kg.

Diagnosis: inflammatory right colon symmetric stenosis and small bowel occlusion.
**Same patient**: Ileocaecal surgical resection:

**Adenocarcinoma with signet ring cell component**: T4N2M0. Signet ring cell infiltration of all colic layers and positive boards of resection

**Mucosa**: important tumor (signet ring cell) infiltration

**Submucosa**: intense tumoral infiltration

**Muscularis**: poor tumoral infiltration

**Rich vascularisation**: neo-angiogenesis
Same patient: Adenocarcinoma with signet ring cell T4: the signet ring cells infiltrate all layer of the colic wall and disorganize its structure. The enhancement of the inner layer corresponds of the tumor and its vessels.
42 years-old man with CD from 15 years. Rectosigmoiditis and anal fistula, difficult medical treatment. Transverse colostomy of discharge.

Severe rectosigmoiditis with submucosal edema in hypersignal T2, important enhancement of all layers of rectosigmoid and hypersignal in Diffusion. Complex anal fistula at 10h.

Same patient 5 months later. Severe rectosigmoiditis and anal lesion. Small bowel occlusion through the transverse colostomy.
Same patient. Small bowel occlusion through the transvers colostomy.

MRI 10/2008

The aspect of the ileitis in the beginning of the rectosigmoiditis.

Small bowel occlusion due to an ileum inflammatory symmetric stenosis with progressive beginning, infiltration of adjoining fat.

MRI 03/2009

Patient 3
Small bowel occlusion due to an ileum inflammatory circumferential stenosis. Important infiltration in the periileal fat. Little abscess. Ileitis with intense enhancement of the inner layer.

Ileo-colic total resection: adenocarcinoma with signet ring cell component spread out from ileum to the rectosigmoid and the anus, except the transverse colon. T4 N2 M1
Same patient: Adenocarcinoma with signet ring cell T4 spread out from ileum to the rectosigmoid and the anus, except the transverse colon. The signet ring cells infiltrate all the layers, the inner layer is thin, the muscularis is very infiltrated and thick.
31 years old female, CD from 14 years. Adenocarcinoma with signet ring cell right-sided colon 3 years ago diagnosed with small bowel occlusion and perforation.

Recidivism of the colic adenocarcinoma with signet ring cell in the transverse colon. Symmetric stenosis without mass. Intense enhancement of the inner layer without ulceration or abscess.
Same patient: Recidivism of the colic adenocarcinoma with signet ring cell in the transverse colon: intense tumor infiltration of the mucosa and submucosa corresponding of the enhancement.
25 years old man with CD from 11 years, PSC and liver transplantation 3 years ago. Colic adenocarcinoma with signet ring cell component one year ago (2005). Colectomy. Occlusion and rectitis.

MRI: rectitis responsible for small bowel occlusion, resistant to medical treatment. The submucosa is in hypersignal on T2 and with important enhancement after Gadolinium IV. Important local fat infiltration and perirectal ganglia.

Rectoscopy showed local recidive, and a stent was positioned. The occlusion was not resolved and the tumor progressed.
Same patient: Adenocarcinoma with signet ring cells of the right colon T4 (scanner not available), the tumor infiltration is intense in the muscosa and submucosa.
Patient 6

01/2001 CT

38 years-old man, UC from 14 years. Occlusion and difficult medical treatment.

Rectocolitis, enhancement of the inner colic layer without ulceration. Spiculation in the pericolic fat. Ascites.

Diagnosis: severe rectitis and left-sided colitis

Surgery: tumor infiltration of colon, aorta, pelvis, peritoneal carcinomatosis.

Histology: Adenocarcinoma with signet ring cell component T4N2M1
There were 3 patients with CD and concomitant small bowel cancer.

Two patients had ileocaecal affect and one had only small bowel affect.

Two SBC were in the ileum and one was in the duodenum.

2 adenocarcinomas and 1 adenocarcinoma with signet ring cell component.

All SBC presented with occlusion during a disease flare.
43 years old man with CD from 4 years. Small bowel chronic occlusion from 3 months resistant to all medical treatment.

Entero CT at diagnosis, CD with ileitis and inflammatory stenosis of the ileum.

CT: Small bowel occlusion with ileitis and long inflammatory stenosis. Ascites and peritoneal fat infiltration.
Occlusion was not resolved with medical treatment and the patient was operated: ileocolic resection.

**Diagnosis:** *Focal Adenocarcinoma with signet ring cell* component of the ileum on 4 cm of length: T3 N1 M0. The tumor is not macroscopically visible. Corresponds probably to a focal thickening of the ileum stenosis.

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**Malignant stenosis ileum**

- **Mucosa:** intense tumoral infiltration
- **Submucosa:** tumoral infiltration
- **Muscularis:** very poor tumoral infiltration

The tumor is impossible to locate in the long inflammatory stenosis.
The colorectal and small bowel malignancy in IBD are well known.

We confirmed in our series the higher percentage of mucinous and signet ring cell types, the younger age at diagnosis and the relationship with the anatomic location of IBD and cancer.

Most of our patients had Crohn’s Disease.

All adenocarcinomas with signet ring cells presented with occlusion and the preoperative imaging diagnosis was benign inflammatory stenosis.

We could not identify the population of IBD followed in our department: the key words « IBD », « UC » and « Crohn » given too much results. The prevalence and the incidence were not calculated.
The diagnosis of colic or small bowel adenocarcinoma with signet ring cell component is difficult to establish only on imaging findings.

Its appearance is not usual and reminds the stomach linitis.

The signet ring cells infiltrate the colic or small bowel wall without any mass or asymmetric aspect.

The tumor borders were impossible to find even peroperative by surgeon because the signet ring cells do not modify the aspect of the wall.

The wall is rigid and thickened. There was intense enhancement of the inner layer mimicking an inflammatory benign stenosis.

Ascitis and infiltration of peritoneal fat were frequent.
We tried to explain these similarity by *radio-pathologic correlation* in Crohn’s ileitis and small bowel adenocarcinoma with signet ring cell.

We used one patient with CD and malignant stenosis and one patient with CD and inflammatory stenosis, both with small bowel occlusion.
21 years old man, Crohn’s disease
Operated for a inflammatory ileum stenosis

Mucosa: enhancement++

Submucosa and Muscularis: hypodense
important development of the muscularis in this case: impossible de separate these layers on CT.

Mucosa and submucosa: enhancement
=important tumor infiltration

Muscularis: poor enhancement = less important tumor infiltration

43 years old man with CD from 4 years. Small bowel chronic occlusion from 3 months resistant to all medical treatment. Adenocarcinoma with signet ring cell component.
Muscularis

Submucosa: important thickening of muscularis mucosa

Mucosa

Mucosa: intense tumor infiltration

Submucosa: tumor infiltration

Muscularis: poor tumor infiltration

Inflammatory ileum stenosis, CD

Malignant stenosis, CD
Less important tumor infiltration

Tumor infiltration of mucosa and submucosa are accompanied with intense inflammatory infiltration and edema. Richer vascularisation than an inflammatory Crohn’s stenosis: neoangiogenesis

Malignant stenosis: signet ring cell, CD

Muscularis

Submucosa

Mucosa
The intense enhancement of the inner layer in ADC with **signet ring cells** corresponds to the tumor infiltration and NEOANGIOGENESIS.

But the difference is difficult to highlight: this patient had a long inflammatory stenosis and **focal ADC with signet ring cells**: the tumor cannot be located on CT.

*Stenosis on different levels in the same patient: ileitis with local adenocarcinoma*
When the tumor infiltrates totally the colic wall (advanced T4) the enhancement and edema were intense without stratification.
The intestinal tract malignancy is a cause of death in longstanding and severe IBD.

Risk factors are well known. Most important are duration of IBD and anatomic extent.

Most of these cancers have similar imaging presentation to usual small bowel and colorectal cancers.

Large percentage (about 30%) of intestinal tract malignancy in IBD are mucinous adenocarcinoma or adenocarcinoma with signet ring component.

The adenocarcinoma with signet ring component presents as a circular symmetric stenosis and mimics a disease flare with inflammatory stenosis.

Malignant stenosis must be suspected when a patient with IBD and risk factors presents an occlusion by stenosis resistant to medical treatment.
Imaging features of ADC with signet ring cells in IBD versus inflammatory stenosis

The intense enhancement of the inner colic/small bowel layer > 5mm: tumor infiltration and neo angiogenesis of the mucosa and submucosa, visible on 70” after contrast IV injection on CT.

The intense enhancement of all the colic/small bowel without stratification wall when the tumor infiltrates the muscularis: advanced T4.
Abrievations

• IBD: inflammatory bowel disease
• CCR: colorectal cancer
• SBC: small bowel cancer
• CD: Crohn’s disease
• UC: ulcerative colitis
• PSC: primary sclerosing cholangitis
• ADC: adenocarcinoma
References


17. Palascak-Juif, V., et al., *Small bowel adenocarcinoma in patients with Crohn’s disease compared with small bowel adenocarcinoma de novo*. Inflamm
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